Complete Summary

GUIDELINE TITLE

Clinical practice guideline (second edition) for the diagnosis, treatment, and management of reflex sympathetic dystrophy/complex regional pain syndrome (RSD/CRPS).

BIBLIOGRAPHIC SOURCE(S)

Reflex Sympathetic Dystrophy Syndrome Association (RSDSA). Clinical practice guidelines (second edition) for the diagnosis, treatment, and management of reflex sympathetic dystrophy/complex regional pain syndrome (RSD/CRPS). Milford (CT): Reflex Sympathetic Dystrophy Syndrome Association (RSDSA); 2002 Feb. 46 p. [47 references]

GUIDELINE STATUS

This is the current release of the guideline

COMPLETE SUMMARY CONTENT

SCOPE

DISEASE/CONDITION(S)

Reflex sympathetic dystrophy syndrome, also known as:

- Complex regional pain syndrome (CRPS)
- Causalgia (minor or major)
- Sudeck´s atrophy
- Post traumatic dystrophy (minor or major)
- Shoulder hand syndrome
- Reflex neurovascular dystrophy

To facilitate communication and understanding of the full spectrum of signs and symptoms, the designation reflex sympathetic dystrophy syndrome/complex

regional pain syndrome (RSD/CRPS) is generally used throughout these practice guidelines.

GUIDELINE CATEGORY

Diagnosis
Management
Treatment

CLINICAL SPECIALTY

Anesthesiology
Internal Medicine
Neurological Surgery
Neurology
Physical Medicine and Rehabilitation

INTENDED USERS

Advanced Practice Nurses
Health Care Providers
Health Plans
Managed Care Organizations
Occupational Therapists
Patients
Physical Therapists
Physician Assistants
Physicians
Psychologists/Non-physician Behavioral Health Clinicians

GUIDELINE OBJECTIVE(S)

To provide full, unbiased information that enables patients and third-party persons to make informed treatment choices about reflex sympathetic dystrophy syndrome/complex regional pain syndrome (RSD/CRPS)

TARGET POPULATION

Individuals with signs or symptoms suggestive of RSD/CRPS

INTERVENTIONS AND PRACTICES CONSIDERED

Diagnosis

1. Assessment of clinical features
    a. Pain
    b. Skin changes
    c. Swelling
    d. Movement disorder
    e. Spreading symptoms
    f. Bone changes

    g. Duration
2. Laboratory diagnostic aids
    a. Thermogram
    b. Bone scans
    c. Sympathetic blocks
    d. X-rays, electromyography (EMG), nerve conduction studies, computed axial tomography (CAT), and magnetic resonance imaging (MRI) studies

Management/Treatment

1. Patient education
2. Written treatment protocols
3. Psychosocial modalities
    a. Pain-coping techniques
    b. Relaxation techniques
    c. Biofeedback
    d. Self-hypnosis
4. Sequential drug trials/medication for chronic pain control
    a. Nonsteroidal anti-inflammatory agents (e.g., aspirin, ibuprofen, naproxen*, indomethacin, etc.)
    b. Agents acting on the central nervous system by an atypical mechanism (e.g., tramadol)
    c. Anti-depressants (e.g., amitriptyline, doxepin, nortriptyline, trazodone, etc)
    d. Oral lidocaine (mexilitine - somewhat experimental)
    e. Anti-convulsants (e.g., carbamazepine, gabapentin)
    f. Oral opioids (e.g., narcotics with names such as Darvon, Vicodin, Loratab, Percocet, morphine, codeine, etc)
    g. Clonidine patch
    h. Klonopin (clonazepam)
    i. Baclofen
    j. Capsaicin cream
5. Physical and occupational therapy
    a. Weight-bearing versus non-weight-bearing exercises
    b. Hydrotherapy
    c. Pressure massage
    d. Moist heat applications
    e. Use of a transcutaneous electronic nerve stimulation (TENS) unit
    f. Pool therapy
6. Sympathetic blocks
    a. Upper extremity, stellate ganglia block (SGB)
    b. Lower extremity, lumbar sympathetic block (LSB)
    c. Local injection of muscle trigger-points
    d. Phentolamine test
    e. Epidural block
    f. Intravenous tourniquet technique
7. Sympathectomy
    a. Traditional approach
    b. Laparoscopic approach
8. Placebo effect for assessing treatment effectiveness
9. Spinal cord stimulation

10. Morphine pump
    a. Implantation of a morphine pump for the spinal infusion of morphine
    b. Implantation of a morphine pump for the spinal infusion of baclofen

*Note from the National Guideline Clearinghouse (NGC): On December 23, 2004, the FDA issued a public health advisory concerning the use of non-steroidal anti-inflammatory drug products (NSAIDs) including the COX-2 selective agents Celebrex (celecoxib), Bextra (valdecoxib), and a non-selective NSAID, naproxen (sold as Aleve, Naprosyn, and other trade name and generic products). See the FDA Web site for more information.

MAJOR OUTCOMES CONSIDERED

- Physical functioning
- Pain
- Health-related quality of life
- Sensitivity and specificity of diagnostic tests

METHODOLOGY

METHODS USED TO COLLECT/SELECT EVIDENCE

Searches of Electronic Databases

DESCRIPTION OF METHODS USED TO COLLECT/SELECT THE EVIDENCE

Not stated

NUMBER OF SOURCE DOCUMENTS

Not stated

METHODS USED TO ASSESS THE QUALITY AND STRENGTH OF THE EVIDENCE

Subjective Review

RATING SCHEME FOR THE STRENGTH OF THE EVIDENCE

Not applicable

METHODS USED TO ANALYZE THE EVIDENCE

Review

DESCRIPTION OF THE METHODS USED TO ANALYZE THE EVIDENCE

Not stated

METHODS USED TO FORMULATE THE RECOMMENDATIONS

Not stated

RATING SCHEME FOR THE STRENGTH OF THE RECOMMENDATIONS

Not stated

COST ANALYSIS

A formal cost analysis was not performed and published cost analyses were not reviewed.

METHOD OF GUIDELINE VALIDATION

External Peer Review
Internal Peer Review

DESCRIPTION OF METHOD OF GUIDELINE VALIDATION

The Reflex Sympathetic Dystrophy Syndrome Association´s (RSDSA) Scientific Advisory Committee, as well as external medical experts, reviewed and commented on the guidelines.

RECOMMENDATIONS

MAJOR RECOMMENDATIONS

Diagnosis of Reflex Sympathetic Dystrophy/Complex Regional Pain Syndrome (RSD/CRPS)

The diagnosis of RSD/CRPS can be made in the context of a history of trauma to the affected area associated with pain that is disproportionate to the inciting event plus one or more of the following:

- Abnormal function of the sympathetic nervous system
- Swelling
- Movement disorder
- Changes in tissue growth (dystrophy and atrophy)

Thus patients do not have to meet all of the clinical manifestations listed above to make the diagnosis of RSD/CRPS. The new complex regional pain syndrome (CRPS) classification system acknowledges this fact by stating that some patients with RSD/CRPS may have a third type of CRPS by categorizing it as "otherwise not specified". There seems to be a small group of patients whose pain following trauma resolves over time, leaving the patient with a movement disorder. The RSD/CRPS may exceed both the magnitude and duration of symptoms expected from the normal healing process expected from the inciting event. Similarly, the diagnosis is precluded by the existence of known pathology that can be explained by the observed symptoms and degree of pain. There are "grades" of this

syndrome described in the literature with symptoms ranging from minor to severe.

Clinical Features of RSD/CRPS

1. Pain

   The hallmark of RSD/CRPS is pain and mobility problems out of proportion to those expected from the initial injury. The first and primary complaint occurring in one or more extremities is described as severe, constant, burning and/or deep aching pain. All tactile stimulation of the skin (e.g., wearing clothing, a light breeze) may be perceived as painful (allodynia). Repetitive tactile stimulation (e.g., tapping on the skin) may cause increasing pain with each tap and when the repetitive stimulation stops, there may be a prolonged after-sensation of pain (hyperpathia). There may be diffuse tenderness or point-tender spots in the muscles of the affected region due to small muscle spasms called muscle trigger points (myofascial pain syndrome). There may be spontaneous sharp jabs of pain in the affected region that seem to come from nowhere (paroxysmal dysesthesias and lancinating pains).

2. Skin changes

   Skin may appear shiny (dystrophy-atrophy), dry or scaly. Hair may initially grow coarse and then thin. Nails in the affected extremity may be more brittle, grow faster and then slower. Faster growing nails is almost proof that the patient has RSD/CRPS. RSD/CRPS is associated with a variety of skin disorders including rashes, ulcers and pustules. Although extremely rare, some patients have required amputation of an extremity due to life-threatening reoccurring infections of the skin. Abnormal sympathetic (vasomotor changes) activity may be associated with skin that is either warm or cold to touch. The patient may perceive sensations of warmth or coolness in the affected limb without even touching it (vasomotor changes). The skin may show increased sweating (sudomotor changes) or increased chilling of the skin with goose flesh (pilomotor changes). Changes in skin color can range from a white mottled appearance to a red or blue appearance. Changes in skin color (and pain) can be triggered by changes in the room temperature, especially cold environments. However, many of these changes occur without any apparent provocation. Patients describe their disease as though it had a mind of its own.

3. Swelling

   Pitting or hard (brawny) edema is usually diffuse and localized to the painful and tender region. If the edema is sharply demarcated on the surface of the skin along a line, it is almost proof that the patient has RSD/CRPS. However, some patients will show a sharply demarcated edema because they tie a band around the extremity for comfort. Therefore, one has to be certain that the sharply demarcated edema is not due to a previously wrapped bandage around the extremity.

4. Movement disorder

Patients with RSD/CRPS have difficulty moving because they hurt when they move. In addition, there seems to be a direct inhibitory effect of RSD/CRPS on muscle contraction. Patients describe difficulty in initiating movement, as though they have "stiff" joints. This phenomena of stiffness is most noticeable to some patients after a sympathetic nerve block when the stiffness may disappear. Decreased mobilization of extremities can lead to wasting of muscles (disuse atrophy). Some patients have little pain due to RSD/CRPS but, instead, they have a great deal of stiffness and difficulty initiating movement. Tremors and involuntary severe jerking of extremities may be present. Psychological stress may exacerbate these symptoms. Sudden onset of muscle cramps (spasms) can be severe and completely incapacitating. Some patients describe a slow "drawing up of muscles" in the extremity due to increased muscle tone leaving the hand-fingers or foot-toes in a fixed position (dystonia).

5. Spreading

Initially, RSD/CRPS symptoms are generally localized to the site of injury. As time progresses, the pain and symptoms tend to become more diffuse. Typically, the disorder starts in an extremity. However, the pain may occur in the trunk or side of the face. On the other hand, the disorder may start in the distal extremity and spread to the trunk and face. At this stage of the disorder, an entire quadrant of the body may be involved. (Note: A small percentage of patients have developed generalized RSD affecting the entire body.)

Three patterns of spreading symptoms have been described: (a) a "continuity type" of spread where the symptoms spread upward from the initial site, such as from the hand to the shoulder, (b) a "mirror-image type" where the spread was to the opposite limb, and (c) an "independent type" where symptoms spread to a separate, distant region of the body. The independent type of spread may be related to a second trauma.

6. Bone changes

Radiological exam (X-rays) may show wasting of bone (patchy osteoporosis) or a bone scan may show increased or decreased uptake of a certain radioactive substance (technetium 99m) in bones after intravenous injection.

7. Duration

The duration of RSD/CRPS varies. In mild cases it may last for weeks followed by remission. In many cases the pain continues for years and, in some cases, indefinitely. Some patients experience periods of remission and exacerbation. Periods of remission may last for weeks, months, or years.

Staging

The staging of RSD/CRPS is a concept that is dying. The course of the disease seems to be so unpredictable among various patients that staging is not helpful in its treatment. Not all of the clinical features listed below for the various stages of

RSD/CRPS may be present. The speed of progression varies greatly in different individuals. Stage I and II symptoms begin to appear within a year. Some patients do not progress to Stage III. Furthermore, some of the early symptoms (Stage I and II) may fade as the disease progresses to Stage III.

Stage I

1. Onset of severe pain limited to the site of injury
2. Increased sensitivity of skin to touch and light pressure (hyperasthesia)
3. Localized swelling
4. Muscle cramps
5. Stiffness and limited mobility
6. At onset, skin is usually warm, red and dry and then it may change to a blue (cyanotic) in appearance and become cold and sweaty.
7. Increased sweating (hyperhydrosis)
8. In mild cases this stage lasts a few weeks, then subsides spontaneously or responds rapidly to treatment.

Stage II

1. Pain becomes even more severe and more diffuse
2. Swelling tends to spread and it may change from a soft to hard (brawny) type
3. Hair may become coarse then scant, nails may grow faster then grow slower and become brittle, cracked and heavily grooved
4. Spotty wasting of bone (osteoporosis) occurs early but may become severe and diffuse
5. Muscle wasting begins

Stage III

1. Marked wasting of tissue (atrophic) eventually become irreversible
2. For many patients the pain becomes intractable and may involve the entire limb.

Laboratory Diagnostic Aids

There is no laboratory test that can stand alone as proof of RSD/CRPS. However, there are a couple of tests (thermogram and bone scan) which can be useful in providing evidence for RSD/CRPS. (refer to the "legal issues" and "laboratory diagnostic aids" sections of the original guideline for details on thermogram use and bone scans).

Treatment

The single most important modality for treating the patient with reflex sympathetic dystrophy/complex regional pain syndrome (RSD/CRPS) is education. The informed consent process should be the focus of education. The physician defines the potential benefits, risks, alternatives (and costs). From the start, the therapeutic goals must be defined and accepted by the patient:

- Educate about therapeutic goals

- Encourage normal use of the limb (physical therapy)
- Minimize pain
- Determine the contribution of the sympathetic nervous system to the patient´s pain.

The cornerstone in the treatment of RSD/CRPS is normal use of the affected part as much as possible. Therefore, all modalities of therapy (drugs, nerve blocks, TENS [transcutaneous electronic nerve stimulation], physical therapy, etc.) are employed to facilitate movement of the affected region of the body. Although physical therapy is an important treatment modality, significant misuse and overuse of this modality may occur. Often the physical therapist will treat the patient with RSD/CRPS the same as a stroke or a nerve plexus injury, (which will fail due to extreme pain with passive manipulation).

The primary goal of the physical therapist should be to teach the patient how to use their affected body part through activities of daily living. Swimming pool exercises are very helpful, especially for RSD/CRPS of the lower extremity where weight-bearing can be problematic. The goal of physical therapy should be to create independence from the health care system in the shortest period. Learning that "to hurt is not to harm" is difficult, but it is essential to avoid reinjury.

Learning the non-protective nature of pain due to RSD/CRPS takes time. For patients who are significantly impaired in their ability to mobilize their extremity, it is urgent to offer the patient the opportunity to determine the contribution of their sympathetic nervous system to their pain. This is accomplished by a sympathetic nerve block to the affected extremity. Future therapeutic options for the patient will depend on whether their pain is determined to be sympathetically maintained pain (SMP) or sympathetically independent pain (SIP). Published reports suggest that the best response to sympathetic blocks will occur if the blocks are given within 6 months of the injury.

The "LET´S TRY THIS NOW" approach is to be deplored because it indicates that the physician has not defined a strategy to achieve specific therapeutic goals in the shortest period of time. It also adds to the confusion, frustration, anxiety and depression of the patient, which may intensify the patient's pain and adversely effect the doctor-patient relationship.

1. Establish a Written Treatment Protocol

   Initiate the safest, simplest, and most cost-effective therapies first. (Refer to Figure 2 in the original guideline for details). If the patient fails to progress in mobilizing the extremity, it is essential to offer the patient a series of three sympathetic blocks immediately. The purpose of the sympathetic blocks is three-fold: (a) to treat, (b) to diagnose if the pain is sympathetically maintained, and (c) to provide prognostic information. The sympathetic block provides a prognostic indicator if sympathectomy or other treatment modalities would be the next appropriate step (sympathetic blocks are described in more detail in the section headed "Sympathetic Blocks").

   After the physician has completed a defined course of treatment (such as a series of 3 to 6 sympathetic blocks), it would be helpful to prepare an update report that would document the patient´s response to the course of

treatment. The report should reflect a basis for further treatment and it should address future rehabilitation needs. Sharing a copy of the update report with the patient will help ensure that all parties (e.g., other medical providers, insurance carriers, case managers, attorneys, etc) are kept informed. Sharing the report with the patient helps keep the patient and physician focused on achieving appropriate therapeutic goals.

An update report should address five areas of care:

1. Procedures (such as nerve blocks)
2. Medications
3. Physical/occupational therapy
4. Psychosocial issues
5. New laboratory tests or consults

2. Psychosocial Modalities

Psychosocial modalities must be considered in all patients with RSD/CRPS. Patients with severe, advanced stage RSD/CRPS usually undergo a psychosocial evaluation during the series of sympathetic blocks or prior to offering the patient more invasive treatments.

In some cases, a formal psychosocial evaluation should be initiated much earlier in the course of treatment. For example, children with RSD/CRPS may require a thorough evaluation to determine the family support structure and the coping mechanisms needed by the family for optimal rehabilitation of the child. The psychosocial evaluation should always be done by an expert in chronic pain and should always include an assessment of pain coping skills and drug abuse potential. Stress is a known cause of exacerbation of this disease, making emergency treatment more necessary. A lot of memorials sent to the Reflex Sympathetic Dystrophy Syndrome Association (RSDSA) Research Fund are the result of suicides! The potential for committing suicide needs to be assessed. The patient may need to participate in a formal pain management program as an outpatient or an inpatient. Chronic pain patients referred for a psychosocial evaluation tend to be defensive. An MMPI (Minnesota Multiphasic Personality Inventory) or other psychological test can help identify the psychosocial problems. Patients must be properly motivated to improve their coping skills; otherwise, application of these psychosocial modalities is a waste of time. Relaxation techniques, such as breathing exercises, as well as biofeedback and self-hypnosis, may be appropriate treatment modalities for some patients.

3. Sequential Drug Trials

Try to initiate sequential trials for each modality of therapy. The application of multiple therapies at the same time, a shotgun approach, makes it almost impossible to evaluate and optimize an individual therapy for safety and efficacy. Patients must be advised that the optimal dose for medications varies greatly among patients. Therefore, it is usually necessary to gradually increase the dose of their medication to the point of significant toxicity in order to determine optimal dose. The dose is then reduced to the next lower level. Thus, it is important for the patient to become familiar with all of the potential side effects of a medication before trying it. Sequential trials with

many different drugs may be required to determine the best medication for the patient. Medications are generally prescribed according to the following characteristics of the pain:

- Constant pain
- Pain causing sleep problems
- Inflammatory pain or pain due to recent tissue injury
- Spontaneous jabs (paroxysmal dysesthesias and lancinating pain)
- Sympathetically maintained pain (SMP)
- Muscle cramps

Medications used to treat chronic pain

"Off-labeling" prescribing means that the U.S. Food and Drug Administration (FDA) approved the medication for one purpose but it is used by physicians for another purpose. For example, aspirin is a pain medication, but it can also be used to decrease the risk of a heart attack by inhibiting the aggregation of platelets. Off-label prescribing is a common practice in treating various chronic pain problems. Some of these drugs have been proven to be effective in decreasing pain due to nerve injury (neuropathic pain) in well-controlled clinical trials. Since RSD/CRPS is believed to be caused by nerve injury (neuropathic pain), these drugs are used to treat this condition as well. The patient should consider weaning themselves from these various medications periodically with the treating physician´s knowledge to determine for themselves that the medication is actually helping to alleviate their symptoms. Some medications need to be weaned slowly (such as narcotics or baclofen) to minimize withdrawal symptoms.

Medications commonly used to treat RSD/CRPS based on the type of pain include:

For constant pain associated with inflammation:
Nonsteroidal anti-inflammatory agents (e.g., aspirin, ibuprofen, naproxen*, indomethacin, etc.)

*Note from the National Guideline Clearinghouse (NGC):
On December 23, 2004, the FDA issued a public health advisory concerning the use of non-steroidal anti-inflammatory drug products (NSAIDs) including the COX-2 selective agents Celebrex (celecoxib), Bextra (valdecoxib), and a non-selective NSAID, naproxen (sold as Aleve, Naprosyn, and other trade name and generic products). See the FDA Web site for more information.

For constant pain not caused by inflammation:
Agents acting on the central nervous system by an atypical mechanism (e.g., tramadol)

For constant pain or spontaneous (paroxysmal) jabs and sleep disturbances:
Anti-depressants (e.g. amitriptyline, doxepin, nortriptyline,

trazodone, etc)
Oral lidocaine (mexilitine - somewhat experimental)

For spontaneous (paroxysmal) jabs:
Anti-convulsants (e.g. carbamazepine, gabapentin may relieve constant pain as well)

For widespread, severe RSD/CRPS pain, refractory to less aggressive therapies:
Oral opioids. The use of opioids (e.g., narcotics with names such as Darvon, Vicodin, Loratab, Percocet, morphine, codeine, etc) to treat RSD/CRPS is debated and there are potential hazards. In order to ensure appropriate informed consent, it is recommended that the patient sign a doctor-patient "contract" (refer to the "Opioid Treatment Protocol" in the original guideline for details).

Patients may require immediate and adequate pain relief. In some cases it may take time to transpire from the time of the patients first visit to the time of adequate treatment. In all probability, the pain and degenerative cycle would progress. Since the abuse potential is minimal when narcotics are used for severe pain, practitioners should not withhold narcotic treatment if the patient demonstrates pain relief with this medication.

For the treatment of sympathetically maintained pain (SMP):
Clonidine patch

For muscle cramps (spasms and dystonia):
Klonopin (clonazepam)
Baclofen

For localized pain related to nerve injury:
Capsaicin cream. (This medication is applied to the skin and behaves like hot peppers. The effectiveness of capsaicin cream in the treatment of RSD/CRPS has not been determined).

4. Physical and Occupational Therapy

Patients need to be educated on how to use their affected body part through activities of daily living. For example, for lower extremity RSD/CRPS, patients may need to be taught weight bearing versus non-weight-bearing exercises. Hydrotherapy is usually medically necessary for muscle (myofascial) pain and spasms. Application of pressure (massage) and/or moist heat applications can sometimes relieve severe muscle cramps. The physical therapist can also teach the patient how to use a TENS (transcutaneous electronic nerve stimulation) unit, a noninvasive electrical device that stimulates the surface of the skin. Pool therapy can be very effective for improving mobility.

5. Sympathetic Blocks

There are three reasons to consider sympathetic blockade to facilitate the management of RSD/CRPS.

- The sympathetic block may provide a permanent cure or partial remission of RSD/CRPS.
- By selectively blocking the sympathetic nervous system the patient (and physician) will gain further diagnostic information about what is causing the pain. The sympathetic block helps determine what portion of the patient´s pain is being caused by malfunction of their sympathetic nervous system.
- The patient´s response to a sympathetic block provides prognostic information about the potential merits of other treatments.

The maximum sustained benefit from a series of sympathetic blocks is usually apparent after a series of 3 to 6 blocks. Even if the original site is unresponsive to sympathetic blockade, future exacerbation of RSD/CRPS symptoms at the same site or at a distant site may be responsive to 1 to 3 sympathetic blocks. (Refer to the treatment section of the original guideline for details and rationale for sympathetic blockade) THE GOAL IS ALWAYS TO TREAT BUT DON´T OVER TREAT.

6. Sympathectomy

If there is a significant decrease in pain following the sympathetic block, the patient is said to have sympathetically maintained pain (SMP). If there is not a significant decrease in pain, the patient has sympathetically independent pain (SIP). Only patients with sympathetically maintained pain (SMP) should be considered for a sympathectomy.

Recently, laparoscopic sympathectomy has been developed for sympathectomy of the upper extremity. This technique requires the placement of three small holes temporarily in the side of the chest wall while the patient is under general anesthesia. For the lower extremity, the patient has the choice of dissolving (destroying) the sympathetic nerves with phenol injected through a needle while the patient is awake (percutaneous phenol sympathetic neurolysis) or a surgical sympathectomy under general anesthesia. Other techniques for sympathectomy have also been used. The patient must be informed of the pros and cons of each approach.

Post-sympathectomy pain (neuralgia) is a potential complication of all types of sympathectomy. Post-sympathectomy pain is typically proximal to the original pain (e.g. proximal means that the pain may appear for the first time in the groin or buttock region for sympathectomy of the lower extremity and pain in the chest wall region for sympathectomy of the upper extremity). Patients may think that their RSD/CRPS has spread to a new region after sympathectomy because the pain feels similar to their original RSD/CRPS pain. The post-sympathectomy pain usually resolves on its own or with 1 to 3 sympathetic blocks. Thus, for some patients, sympathectomy may be a two-step procedure; destruction of sympathetic nerves followed by a sympathetic block. Data published by the Reflex Sympathetic Dystrophy Syndrome Association (RSDSA) suggests that sympathectomy in properly selected RSD/CRPS patients may provide one of the most effective treatments for

RSD/CRPS. The selection criteria for sympathectomy are critical in achieving long-term success.

7. Placebo

The placebo effect (decreased pain due to an inactive treatment such as a sugar pill) must be considered in the treatment of RSD/CRPS. Although the figure of 33% is commonly quoted in papers and textbooks as the percentage of people who will respond to a placebo, it is misleading because the "percentage" varies enormously (from close to 0 to 100%) depending on the exact circumstances. Physician and patient must have an understanding about the placebo effect, otherwise the patient is at risk of being over-treated. Recognition of placebo versus specific pain-relieving treatment may be difficult, but there are some distinguishing characteristics:

- The greater the invasiveness of the procedure itself, the greater the placebo effect.
- The greater the expectation for pain relief, the greater will be the placebo effect.
- The placebo tends to be of less duration. For example, close monitoring of the patient's pain for hours and days after each sympathetic nerve block has shown that the pain-reducing effect of the saline (placebo) injection subsides within the first few hours, whereas that of the local anesthetic injection persists for several days.
- The placebo tends to be less reproducible with each successive treatment.

Therefore, it may be of great potential therapeutic value to provide each patient with a series of multiple sympathetic blocks separated by brief intervals (such as one week) simply to determine whether such blocks are effective treatments. (Refer to the treatment section of the original guideline for further details on placebo effect.)

8. Other Types of "Sympathetic Blocks"

A sympathetic blocker (alpha adrenergic antagonist), phentolamine, given intravenously (I.V.), has been advocated as a diagnostic test for sympathetically maintained pain (SMP). However, false negative tests have been reported as high as 43%. Moreover, this approach is somewhat elaborate and requires considerable technician time and expense. The phentolamine test is a diagnostic procedure while a sympathetic block is a diagnostic, prognostic and therapeutic procedure. However, the phentolamine test may be valuable treatment option in the situation where a sympathetic block is not possible or when multiple extremities are involved.

Epidural blocks are less specific for blocking the sympathetic nervous system and, therefore, they are not as useful for diagnostic and prognostic objectives. The infusion of local anesthetic through the epidural catheter may cause temporary weakness in the legs, making walking dangerous. Placement of long-term epidural catheters to treat RSD/CRPS still occurs in practice. Perhaps this is because anesthesiologists are more familiar with the epidural catheter technique than with the selective sympathetic block technique. The

long-term epidural catheter approach is more expensive and patients are placed at more risk for certain rare life-threatening complications, such as infection (epidural abscess). Often, a short (2 to 5 days) hospitalization will be necessary to determine the clinically most appropriate dose of the epidural agent for constant infusion. Dislodgment of the epidural catheter is a relatively common problem. The use of a lumbar sympathetic catheter may provide a more specific sympathetic block than an epidural catheter, but the lumbar sympathetic catheter is more likely to become dislodged during exercise. There is a place for the use of epidural and lumbar sympathetic catheters in the treatment of RSD/CRPS, but the physician should justify these techniques on a case by case basis.

Another technique used to carry out a sympathetic block involves the intravenous injection of sympathetic blocking agents (such as guanethidine, bretylium and clonidine) into an extremity and limiting spread of the agent to the entire body by applying a tourniquet to the extremity. This method requires placing an IV 'catheter' in the painful extremity and may be technically extremely difficult due to severe swelling (edema) of the extremity. The patient may not be able to confirm that they actually received a sympathetic block because the "cue", a warming sensation in the extremity, may not be felt. Furthermore, there is no evidence that this technique is more effective than the usual sympathetic blocks for the diagnosis and treatment of RSD/CRPS. The IV tourniquet technique using a sympathetic blocking agent may be considered as an option for patients who must take blood thinners (anticoagulants), where a stellate ganglia block (SGB) or a lumbar sympathetic block (LSB) may cause major bleeding.

9. Spinal Cord Stimulation

Spinal cord stimulation (SCS) is another method of pain control that works well for some patients with chronic, intractable pain due to RSD/CRPS. SCS uses low intensity, electrical impulses to trigger selected nerve fibers along the spinal cord (dorsal columns), which are believed to stop pain messages from being transferred to the brain. SCS replaces the area of intense pain with a more pleasant tingling sensation called paresthesia. The tingling sensation will remain relatively constant and should not hurt. There is some experimental evidence that SCS may enhance the flow of blood to the affected extremity by blocking the sympathetic nervous system.

A temporary trial, with a temporary electrode, should be performed first before implanting permanent electrode(s). Given that SCS is a relatively invasive, costly procedure and given that RSD/CRPS patients are often desperate and frustrated, a baseline psychosocial evaluation that addresses pain management issues should be considered. Although rare, spinal infection and paralysis are potential complications. The ability to insert the electrode through a small needle has reduced the risk of the procedure and has facilitated the trial with a temporary electrode.

Treating RSD/CRPS with SCS poses unusual clinical and technical problems, as RSD/CRPS tends to be an unpredictable disease from a technical standpoint. The need to focus SCS on the most painful region must be kept in mind, which is more difficult in RSD/CRPS, because the location of the worst

pain may change. Furthermore, the pain from RSD/CRPS may spread to distant parts of the body, requiring multiple, successive implanted stimulators to cover the largest possible area. Therefore, even when RSD/CRPS is limited to one extremity, it is wise to widen stimulation to zones to which the pain might spread.

Because of the risks and high costs of spinal cord stimulation, the treatment is reserved for severely disabled patients. A recent well-controlled study shows that with careful selection of patients and successful test stimulation, SCS is safe, reduces pain, and improves the health-related quality of life in patients with severe RSD/CRPS.

In order to make an informed choice about SCS, the patient and physician should consider the pertinent differences between the internal and external battery systems (refer to the original guideline document for details on comparisons between internal and external battery systems for spinal cord stimulation).

10. Morphine Pump

It is well-recognized that a single injection of morphine into the spinal fluid (within the intrathecal space) produces a selective pain-blocking effect on the spinal cord. This selective effect on the spinal cord spares the patient from many of the serious side effects caused by morphine when it is given orally (such as sedation). Soon after this discovery, enthusiasm developed to implant permanent morphine pumps to treat non-cancer chronic pain, especially after Medicare began to approve this surgical procedure for reimbursement. The implantation of a morphine pump is a relatively invasive and expensive treatment modality. Despite almost 20 years of testing, no scientific evidence has emerged that long-term use of the morphine pump offers an advantage over oral morphine. In fact, many patients with the implanted morphine pump take oral morphine at the same time. The same complications sometimes associated with oral morphine use are also found with the morphine pump, such as development of drug tolerance, nausea, constipation, weight gain, decreased sex appetite (libido), swollen legs (edema), and increased sweating. In addition, malfunction of the pump system (dislodgment of the catheter) can be a significant problem.

A recent study suggests that, with careful selection of patients, the implantation of a morphine pump for the spinal infusion of baclofen may be a valuable means for treating a certain type of muscle cramp called dystonia in patients with RSD/CRPS.

How to Determine the Effectiveness of Treatments for Reflex Sympathetic Dystrophy/Complex Regional Pain Syndrome (RSD/CRPS)

Refer to the original guideline document for details on the natural history of RSD/CRPS, placebo or nonspecific effects of treatment, and specific effects of treatment.

CLINICAL ALGORITHM(S)

The original guideline document contains a treatment protocol clinical algorithm.

EVIDENCE SUPPORTING THE RECOMMENDATIONS

TYPE OF EVIDENCE SUPPORTING THE RECOMMENDATIONS

The type of supporting evidence is not specifically stated for each recommendation.

BENEFITS/HARMS OF IMPLEMENTING THE GUIDELINE RECOMMENDATIONS

POTENTIAL BENEFITS

- Appropriate, timely diagnosis and treatment of reflex sympathetic dystrophy/complex regional pain syndrome (RSD/CRPS)
- If diagnosed early, physicians can use mobilization of the affected extremity (physical therapy) and sympathetic nerve blocks to cure or mitigate the disease. If untreated, RSD/CRPS can become extremely expensive due to permanent deformities and chronic pain.
- Minimization of pain
- Improved quality of life

POTENTIAL HARMS

Oral Opioid Use

There is potential for abuse and/or life-threatening overdoses with the use of oral opioids.

Sympathetic Block

Complications from sympathetic blockade are extremely rare. However, it is always possible for the local anesthetic to be inadvertently injected into a blood vessel or into the spinal fluid. If this should happen, the patient may temporarily become weak and lose consciousness.

Sympathectomy

Post-sympathectomy pain (neuralgia) is a potential complication of all types of sympathectomy. The post-sympathectomy pain usually resolves on its own or with 1 to 3 sympathetic blocks.

Epidural Blocks

The infusion of local anesthetic through the epidural catheter may cause temporary weakness in the legs, making walking dangerous. Dislodgment of the epidural catheter is a relatively common problem. The long-term epidural catheter approach is more expensive and patients are placed at more risk for certain rare life-threatening complications, such as an infection (epidural abscess).

Morphine Pump

Complications sometimes associated with oral morphine use are also found with the morphine pump, such as development of drug tolerance, nausea, constipation, weight gain, decreased sex appetite (libido), swollen legs (edema), and increased sweating. In addition, malfunction of the pump system (dislodgment of the catheter) can be a significant problem.

Spinal Cord Stimulation

Although rare, spinal infection and paralysis are potential complications.

IMPLEMENTATION OF THE GUIDELINE

DESCRIPTION OF IMPLEMENTATION STRATEGY

An implementation strategy was not provided.

IMPLEMENTATION TOOLS

Clinical Algorithm

For information about availability, see the "Availability of Companion Documents" and "Patient Resources" fields below.

INSTITUTE OF MEDICINE (IOM) NATIONAL HEALTHCARE QUALITY REPORT CATEGORIES

IOM CARE NEED

Living with Illness

IOM DOMAIN

Effectiveness
Patient-centeredness

IDENTIFYING INFORMATION AND AVAILABILITY

BIBLIOGRAPHIC SOURCE(S)

Reflex Sympathetic Dystrophy Syndrome Association (RSDSA). Clinical practice guidelines (second edition) for the diagnosis, treatment, and management of reflex sympathetic dystrophy/complex regional pain syndrome (RSD/CRPS). Milford (CT): Reflex Sympathetic Dystrophy Syndrome Association (RSDSA); 2002 Feb. 46 p. [47 references]

ADAPTATION

Not applicable: The guideline was not adapted from another source.

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GUIDELINE DEVELOPER(S)

Reflex Sympathetic Dystrophy Syndrome Association - Private Nonprofit Organization

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GUIDELINE COMMITTEE

Scientific Advisory Committee

COMPOSITION OF GROUP THAT AUTHORED THE GUIDELINE

Editor: Anthony F. Kirkpatrick, MD, PhD (Chair)

Committee Members: Charles Berde, MD, PhD; Wen-hsien Wu, MD, MS; Timothy Lubenow, MD; Robert Jay Schwartzman, MD; Srinivasa Raja, MD; Gustavo C. Roman, MD; Sabine Kost-Byerly, MD; Adam L. Seidner, MD, MPH; Peter Wilson, MD

FINANCIAL DISCLOSURES/CONFLICTS OF INTEREST

Not stated

GUIDELINE STATUS

This is the current release of the guideline

GUIDELINE AVAILABILITY

Electronic copies: Available from the Reflex Sympathetic Dystrophy Syndrome Association (RSDA) Web site.

Print copies: Available from the Reflex Sympathetic Dystrophy Syndrome Association; PO Box 502, Milford, CT 06460; Phone: 203-877-3790; Web site: www.rsds.org

AVAILABILITY OF COMPANION DOCUMENTS

None available

PATIENT RESOURCES

None available

NGC STATUS

This NGC summary was completed by ECRI on December 11, 2002. The information was verified by the guideline developer on January 27, 2003. This summary was updated by ECRI on January 12, 2005 following the release of a public health advisory from the U.S. Food and Drug Administration regarding the use of some non-steroidal anti-inflammatory drug products.

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